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Case Report

Esophageal carcinoma complicating achalasia, 25 years after esophagomyotomy

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Abstract

The development of Esophageal carcinoma with achalasia long after adequate surgical treatment is a rare occurrence which is seldom documented in clinical reports. We present a report of oeseophageal Carcinoma in an 82year old woman with achalasia of the esophagus 25 years after esophagomyotomy. Serial Imaging and Endoscopic examinations revealed the malignant transformation of the achalasia. This emphasizes the need for life long surveillance in the management of cancer oesophagus..

Introduction

The first case of achalasia complicated by esophageal carcinoma was reported in 1872 [1]. Since this initial observation, numerous case reports and retrospective reviews have appeared to seemingly substantiate the validity of this association, with the reported frequency of associated esophageal cancer in subjects with achalasia being as high as 20% [2], although most series have found a lower frequency in the order of 5% [3-7,]Treatment of achalasia, an incurable disorder of esophageal motility, offers at least partial symptomatic relief for most patients [8]. Although this abnormality affects both the esophageal body and the lower esophageal sphincter, therapy is directed only at the sphincter [8]. Some degree of stasis usually persists despite adequate treatment and relief of symptoms. Complications or recurrent symptoms include dysphagia, esophageal reflux, stricture and carcinoma [5]. This case report suggests that long term surveillance of the patient with achalasia is essential even after successful treatment.

Case Report

A. M. is an 82 year old woman who was admitted to the surgical ward via the surgical outpatient clinic with 1year history of dysphagia, associated weight loss and regurgitation. Twenty five years previously , a clinical and radiological diagnosis of esophageal Achalasia was made, with marked retention and esophageal dilation seen on Barium Swallow examination (fig. 1). She subsequently under went esophagomyotomy at University College Hospital, Ibadan. During routine follow up examination 5 years after the operation, Barium Swallow (fig. 2.) showed moderate dilation of the esophagus with trickling of barium into the stomach and some esophageal retention Flexible esophago-gastro-duodenoscopy was carried out which showed cobble stone appearance of lower esophageal mucosa plus food debris. Biopsy taken during this procedure revealed chronic oesophagitis. The patient was lost to follow up until 25 years later with a most recent presentation of abrupt onset of dysphagia, weight-loss and regurgitation. Barium Swallow (fig.3) obtained showed a filling defect on the right lateral lower border of the esophagus suggestive of neoplasm. The proximal esophagus was moderately dilated. Endoscopic examination of the esophagus disclosed esophageal dilation and a stricture at 35cm from the incisor teeth. A biopsy of the lesion revealed squamous cell carcinoma. The patient underwent a Stamm's feeding gastrostomy and was scheduled for esophagectomy, cervical esophagogastrostomy and pyloromyotomy, but died shortly before surgery.
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Fig. 1 Barium swallow (before surgery) showing marked dilatation of the entire esophagus with marked retention of barium

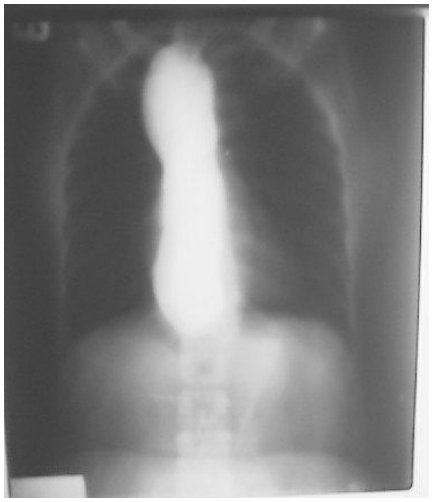


Fig. 2: Barium swallow (5 years post-surgery Moderate dilatation of the oesophagus with retention of barium Barium trickling into the stomach

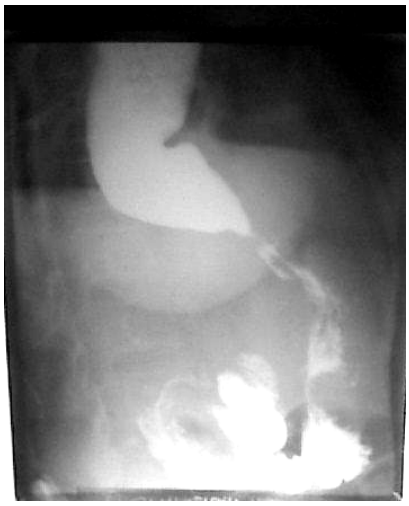
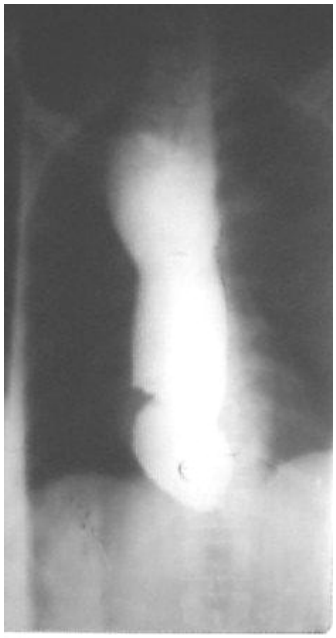


Fig. 3 Barium swallow (25 years post surgery):
Filling defect seen on the R lateral lower border of the oesophageal wall



Discussion

The association of carcinoma of the esophagus with achalasia, first reported by Fagge[1] in 1872, appears to be well established. Subsequent reports have appeared in the literature, and reviewed by several authors [3-7]. The reported incidence varies widely, presumably reflecting disparate methods of study and referral patterns. In their review, Bolivar and Herendeen [8] cited the frequency of occurrence of clinically determined carcinoma in achalasia to be 0-7.7%, but found a higher incidence of 20-29% at autopsy. Wychulis and associates [9] reported on 1,318 patients treated for esophageal achalasia at the Mayo clinic for an average of 13 years per patient. Carcinoma developed in seven of these patients, an incidence almost seven times that in the general population. In Nigeria, Pindiga [10] retrospectively analysed 177 histologically confirmed cases of oesophageal carcinoma seen in the University College Hospital Ibadan, over a period of 30 years. They found out that none was associated with achalasia. The patient in this report presented with oesophageal Carcinoma complicating achalasia and this was probably one of the few cases seen over a period of 36 years. This confirmed the rarity of this lesion.Carcinoma of the esophagus arises at an earlier age in person with achalasia than it does in persons with a normal esophagus[11]. In one large review[12], cancer developed in patients whose average age was 48 years in contrast to an average age of 62 years in the normal population; this is in contrast with this case report presented aged 82 years. Reports of carcinoma in patients younger than aged 40 years

with achalasia are common. In a collected review of the literature [12,] symptoms of achalasia preceded the development of carcinoma by an average of 17 years. Others 9 have found this duration to be considerably longer which is in agreement with the case presented, with a duration of 25 years.

The cause of carcinoma occurring in achalasia is unknown. The commonest view, however, is that chronic irritation resulting from stasis is the major factor. In 1975, Hankins [13] indicated that stasis plays an important role in the development of carcinoma in achalasia. He observed that malignant change was found in the dilated esophageal pouch and not at the stenotic portion of the esophagus. Rake [2] found undetected carcinoma in three of 15 patients who died of achalasia. Diffuse mucosal inflammation and ulceration, corresponding to the height of the column of stagnating material, were present in the majority of patients. Epithelial over growths or papillomas were found in some patients. Rake 2 proposed that these changes eventually resulted in malignant degeneration. Studies [14] show that treatment with Heller's esophagomyotomy and dilation offers good to excellent results in a majority of patients. The results of treatment, however, are usually based on subjective criteria and do not reflect the degree of retention of material in the esophagus [15.] Once megaesophagus has developed stasis is usually incompletely relieved by myotomy. Some authors [5, 8] have expressed the belief that treatment with Heller's myotomy does not protect against the development of carcinoma. Others [6,7,9], holding the opposite view, have advocated early surgical treatment.

Lortat – Jacob [6] observed that in 24 patients with carcinoma associated with achalasia, nine patients had had a prior Heller operation, but the myotomy was inadequate with remaining esophageal stasis and dilatation.

. Manometric and radiologic studies are often lacking. When reported, the duration of symptoms before operation is frequently lengthy, while the interval between operation and diagnosis of carcinoma is brief. This patient differs in that the interval between operations and the diagnosis of cancer is relatively long . Anima [16] documented that long-term and periodic X-ray and endoscopic observation of the disease course is important, and iodine staining is indispensable. Whether treated by operation or not, patients should receive long-term regular endoscopic and histologic surveillance. Although published reports indicate an average of 17years from the onset of symptoms to the development of carcinoma, the time period has a wide range and much shorter intervals have been reported [9,12]. A reasonable surveillance includes endoscopy with multiple random biopsies every three to five years after adequate dilation or surgery could be beneficial [9]

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